

Webber (S. G.)

PROGRESSIVE LOCOMOTOR ATAXY.

Read before the Boston Society for Medical Observation
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This disease was known formerly as *tabes dorsalis*, and as such is well described by Romberg; but the whole of the group of symptoms belonging to it was not fully recognized until Duchenne (of Boulogne) communicated a memoir on that subject to the Society of Medicine of Paris in 1857. This memoir is reproduced in all its essential particulars in his work "L'Electrisation Localisée." Since that date, observers in other places have continued the investigations, and an extensive literature on the subject has been created.

The two cases here related illustrate two different forms of the disease; but in neither do all the symptoms occur which are sometimes met. For a fuller description reference may be made to Trousseau's *Clinique Médicale de l'Hôtel Dieu*, t. ii.; to St. George's Hospital Reports, vol. i.; an article by Dr. J. L. Clarke; or to Reynolds's *System of Medicine*, vol. ii. Dr. Radcliffe, in the last work, sums up the distinctive symptoms as follows:—

"A peculiar gait, arising from want of coördinating motor power in the lower extremities—a gait precipitate and staggering, the legs starting hither and thither in a very disorderly manner, and the heels coming down with a stamp at each step.

"No true paralysis in the lower extremities or elsewhere.

"Characteristic neuralgic pains, erratic, paroxysmal, in the feet and legs chiefly—pains of a boring, throbbing, shooting character, like those caused by a sharp electric shock.

"More or less numbness in the feet and legs chiefly, in all forms of sensibility, excepting that by which differences of temperature are recognized.

"Frequent impairment of sight or hearing, one or both.

"Frequent transitory or permanent strabismus or ptosis, one or both.

"No very obvious paralysis of the bladder or lower bowel.

"No necessary impairment of sexual power.

"No tingling or kindred phenomena.

"No marked impairment of muscular nutrition and irritability.

"No impairment of the mental faculties.

"Occasional injection of the conjunctivæ, with contraction of the pupils."

Dr. Radcliffe takes no notice, in this summary, of the usual order of precedence in the various symptoms. This, as described by Duchenne, is shown in the first of the following cases. The first period is characterized by "paralysis of one or several of the motor nerves of the eye, complicated with paralysis of the optic nerve, and by erratic, boring pains; the second, by the appearance of defects of coördination, and, soon after or simultaneously, by muscular and cutaneous insensibility, generally in the lower limbs, or sometimes in the upper limbs; the third, by generalization of the disease."

CASE I.—Mr. — came to me last February, in accordance with the suggestion of Dr. Knight. He said he wished to know whether electricity would be of any use in restoring his lost power. He seemed to have very little faith in any kind of treatment, and his mental condition was one of great despondency. As he expressed himself, he thought he might as well die of this disease as of any other, if he was going to die at all.

He is about 59 years of age, has been married thirty years and had one child, which is dead. His business has been to construct and fit the wood-work for machinery. His father died of paralysis. He says he has had rheumatism very severely and frequently in every part of his body for the last thirty years. He is not aware of any cardiac trouble. He has many times during the last thirty years strained himself by lifting heavy weights—has at times been laid up for several days on that account; as he said, has had a "crick" in

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his back by turns for several years. But nothing of this kind had occurred for months previous to his paralysis.

For many years he has had attacks of numbness in his right thumb, so that he could not write, especially in winter. For this he had rubbed his thumb with alcohol. If the attack was very severe, he might have only one during the winter; if less severe, he might have more.

Three or four years ago, his eyes were affected, on which account he consulted Dr. Williams, who told him he had amaurosis. The right eye was the worst, and was almost blind. The affection of the eyes came on rather suddenly. Subsequently, the amaurosis improved. He acknowledges no trouble in the head and no vertigo.

About one year ago last January, he noticed, while at work, that he had suddenly lost power over the left leg. He could not, while sitting on his stool, extend his leg. He did not fall down when he tried to walk, but could draw his leg after him. If he stumbled, however, he could not recover his balance. At the same time with the motor paralysis, there was a slight loss of sensation in the same leg. He seems subsequently to have regained considerable control over the muscles of his leg, or else the paralysis was not so complete as he stated, since, when I saw him, it was not very marked.

For several months there was a slow advance in the paralytic symptoms. The paralysis extended to his right leg, and that became worse than the left. It gradually extended up the right side to the right arm and then to the left arm.

Last December, he had paralysis of motion and somewhat of sensation in the right side of his face, and ptosis of the right eyelid. This did not last long. He wet his face in the morning in cold water, and used friction with a coarse salted towel; to this he ascribes the improvement which took place in his facial paralysis.

He thinks that since December he has been losing ground slowly, and that the disease has been gradually advancing.

Now, he has some difficulty in seeing, especially with his right eye, which reacts rather sluggishly under the influence of light. He has no abnormal sensations, as of itching, tingling, formication, &c. He has a sensation as of a cord bound round about the middle of the left arm. This sensation exists nowhere else. He complains of a sensation of cold nearly all the time, especially in the feet, up to the middle of the leg, and to a less degree in the

body, but most marked in the left great toe and along the inside of the foot to the heel, and here it is accompanied with numbness. It exists to a less degree in corresponding parts of the right foot. The numbness also extends partly up the leg.

He balances tolerably well, and with his eyes shut can walk, though with a very unsteady gait. He can stand with his feet together and eyes shut, but sways backwards and forwards more than is natural in health. He thinks he has most difficulty in moving his legs in the morning, after lying a-bed all night.

When walking, he feels as though he must go fast, so as not to fall forwards, so that his feet may keep up with his head.

Sensation is not very good in the back of his head, and he is very sleepy. He feels best when lying down.

His appetite is poor; he is constipated, but is not troubled with flatus.

The examination ceased here, owing to the lateness of the hour, and I told him I would call the next day at his house for the purpose of examining the condition of the various sensations. The next day, excuse was made that his wife was sick, and I have not heard from him since.

CASE II.—O. McG., a laborer, I saw at the City Hospital, in the service of Dr. Blake, who has kindly permitted me to make use of the case to further illustrate the subject. The hospital record states that he was a hard drinker till eight years ago, and since that time has indulged immoderately in whiskey, but has always been well and strong till last May. About one month before that time he was knocked down by a horse, which trod upon his right side. He is not aware of any injury to his head or back, though after the accident he had much pain in his side on breathing.

About May 1st, he first experienced a cold sensation in the left foot, as of a wet stocking drawn over it. In a month, the right foot was affected in a similar way. The sensation gradually extended, and in three months had reached the knees, when he was obliged to give up work.

He had no headache nor any trouble with his head for three months after the trouble in the lower extremities commenced. He then had sharp, darting pains in teeth, face and head, and since that time has felt light-headed.

At present, the sensation of coldness has invaded the whole of the lower extremities and the body below a line passing through the lower ribs. There is also marked impairment of sensibility, which is worst at

the feet and decreases upwards. Reflex action is very much diminished or lost in the feet. Sensation and motion of the upper extremities seem to be impaired. Eyesight but little affected; he sometimes sees specks floating before his eyes. April 1st, the day of entrance into the hospital, in the afternoon, he complained of a cloudiness of vision for the first time. Pupils act naturally. Taste and smell unimpaired.

He is obliged to micturate quite frequently, and sometimes when he feels the desire to do so, he has a passage of urine unwittingly. He never passes urine while asleep.

His gait, when walking, is peculiarly characteristic of this affection; much more so than in the previous case. When attempting to walk, even if supported on each side, his legs are moved about in a very disorderly manner, sometimes sideways, sometimes forwards and backwards, sometimes crossing each other, and the heel comes down with emphasis upon the floor. If his eyes are shut, he cannot stand when the feet are brought together.

He became dissatisfied and homesick, and left the hospital in two days.

Only occasionally, as at the commencement of the attack in the first case, is there real paralysis of motion; the patient is unable to walk, and is perhaps confined to the bed, but can move his limbs with power, though in a disorderly manner and perhaps by jerks, not being able to check them at just the point desired.

The affection with which locomotor ataxy is most likely to be confounded is disease of the cerebellum, especially one of slow growth, as a tumor. But in an affection of the cerebellum there is found vertigo, sometimes vomiting, headache, or perhaps throbbing sensations in the head. The defect in motion is also different. The gait is unsteady rather from lack of power over the muscles, or on account of the vertigo, than from defect of coördinating power. The vision may be affected in both diseases.

The second case is one of the more erratic cases, commencing with the second period, the symptoms of the first period appearing only intercurrently.

It is certainly a long duration for the disease to have dated back thirty years, yet that is the date at which the rheumatic pains were first felt, and according to his description they seemed to resemble the tearing and boring pains of locomotor ataxy more than those of rheumatism. Though not stated in the notes, I believe he complained of the pains having been in his lower

extremities more frequently and more severely than elsewhere. It would also be very unusual that one should have rheumatism repeatedly, and the pains be so severe that their recollection was disagreeable, and yet be aware of no cardiac complication. I ought to say, however, that I did not make any examination, intending to do so while he was in bed next day. The disease is sometimes of very long duration; thus Mr. Carré refers to cases which had lasted 20, 21, 24 and even 28 years.

In the 2d volume of Reynolds's System of Medicine, Dr. C. B. Radcliffe does not recognize the fact that the upper extremities may be involved. Yet in the first case there was partial loss of sensation in the right thumb, occurring before the loss of motion in the lower limbs. Many other cases are on record, one by Duchenne, in which the abnormal condition was perceived in the upper extremity. Also, at *post mortems*, changes have been recognized in the cervical region of the cord. In a *post mortem* by Dr. J. L. Clarke, recorded in the *Lancet*, June 10, 1865, the change was noticed as high as the second cervical nerve.

The change usually found after death, as described by various observers, is degeneration of the posterior columns of the spinal cord, chiefly in the lumbar region, occasionally extending as high as the cervical, rarely confined to the latter region. This degeneration may extend to the neighboring posterior cornua and gray matter; it may even involve the posterior roots. This last is not a constant lesion, but is usually found where there is loss of sensation. "The degeneration is essentially characterized by gradual and progressive atrophy of the nervous elements, and by a gradual and progressive development of the connective tissue which separates them."—Carré, p. 189.

Similar changes have been found in the optic nerves, and also other cranial nerves when these have been affected, but more especially the motor nerves of the eyes.

Various influences have been assigned as the cause of this disease. Comparatively a large number of patients have suffered either from strains of the back or fatigue, or have been exposed to wet and cold during a long period of time. In both the cases herewith reported, there is a history of injury; in the first case often repeated straining, and the first pains appeared at about the same period as the first strain, or thirty years ago. In the second case, there is a history of an injury to the side, and

consequent difficulty of breathing, about a month before the peculiar coldness of the feet was noticed.

Duchenne states briefly—"Some patients accuse sudden or too prolonged cooling, suppression of transpiration; thus, for example, there is a sportsman who often has remained a long time with the feet and legs in the water. Once, the phenomena of this disease have shown themselves after a too prolonged sitz bath. Trousseau mentions hereditary taint, also, as a predisposing cause. This may appear in the first case, where it is stated that the father died of paralysis.

Out of 132 cases referred to by Carré, some of which, however, were not of this disease, 66 have no cause assigned, and no antecedents are given. In 17 there was a previous history of syphilis, in 8 of rheumatism, 6 had been exposed to unusual and frequent fatigue, 23 had been subjected to cold and dampness in various ways—either dwelling in damp houses, sleeping in the open air on damp ground, working in damp places, or were subject to sudden cooling while perspiring freely, and immediately after felt the first signs of the disease. In 9, there had been emotional excitement or long-continued anxiety. In 5, disease of the nervous system had existed among the ancestors. In 5, abuse of wine or venereal excess had been noted.

Topinard, who has collected a very large number of cases, says that among predisposing causes, "there are few so efficacious, when united together, as disappointment, emotion, and bad hygiene." He also looks upon rheumatism as playing a certain rôle in this relation. With regard to dampness, he says:—"But the most common cause is dampness and sudden cooling, since 16 times it is indicated as determining and 15 times as predisposing, that is to say 31 times in all." He concludes the chapter on Etiology:—"Progressive locomotor ataxy is a disease peculiar to adult age; more frequent in males, in those who are addicted to excesses of all kinds, who are exposed to dampness and to great fatigue. Among temperaments, the nervous alone is certainly disposed to a particular form of the disease. Among diatheses, the rheumatic is the only one whose influence is incontestable." In some cases, he thinks it seems to be hereditary.

Eisenmann recognizes essentially the same causes, only concussion of the spinal cord through falls and strains he considers as perhaps causing rather a local disease, and not properly locomotor ataxy.

The dependency in the first case, and the similar feeling of homesickness in the second, are phenomena worthy of notice.

If the disease is considered a derangement of the vaso-motor system, much that is otherwise anomalous ceases to be so. Many of the earlier symptoms are explained by the existence of congestion of the nerve centres. In the first case, there was a sudden loss of power over the left leg. Previous to that, amaurosis had occurred. Both these symptoms had diminished very much in intensity. The numbness of the right thumb had occurred and disappeared again and again. Later, there was paralysis of the right side of the face, which had nearly or quite disappeared when I saw the patient. In other cases, certain symptoms are found occurring and then disappearing, as strabismus in M. Duchenne's case No. 122; also incontinence of urine. Occasionally the amaurosis, which occurs early in the disease, so far disappears that the patient does not think to speak of it without being questioned. Congestion may well be supposed to be the cause of such temporary symptoms.

Another proof of the truth of this theory is given by M. Trousseau, in the congested state of the conjunctiva and contraction of the pupil which he observed in many patients; but the congestion disappeared and the pupils dilated when there was an access of severe pain. M. Duchenne and Dr. Radcliffe have observed the same.

The means used to dissipate the numbness of the thumb and the paralysis of the face might well act by causing reflex changes in the vessels of the cord:—stimulating friction to the parts affected and cold water to the face.

Another fact directly bearing upon this point may be found in Trousseau's Clinique, in an account by Luys of the microscopic appearances. "In following into the grey substance of the fourth ventricle the investigation upon the trunk of the external motor nerve of the eye, up to its point of real origin, a series of large vascular trunks were seen in the tract of the primitive fibres of this nerve, which they probably must have compressed in a notable degree."

The coldness complained of by both these patients is a proof of vaso-motor disturbance. That this coldness is not merely a subjective sensation there is nothing recorded in recent writers to show; but Hasse states that there is a reduction of 1°-2° R. in the part.

One case, at least, is on record where the patient, dying of some acute disease, little

or nothing has been found to account for the ataxic symptoms.

I quote from Topinard the appearance found after death. The patient had had symptoms of locomotor ataxy since thirteen years, and for six months had been unsteady in his gait. Cutaneous sensibility was altered. He died during an attack of variola, in the last part of which he had true muscular paralysis. "At the autopsy, absolutely nothing was found to the naked eye in the encephalon, cerebellum, the roots or columns of the spinal cord. The whole cord was very much injected; the optic nerves were grey, semi-transparent, softened between the papillæ and the corpora geniculata exclusive. One of the common motor nerves was flattened, diminished in volume, but was not grey. The microscopic examination found the posterior columns and roots healthy, and the usual alteration of the optic nerves. Having myself compared a transverse section of the lumbar enlargement of this patient with a similar section from the ataxic patient of whom we had made the autopsy fifteen days before in the service of M. Trousseau, I have been able to testify to that integrity. However, in more closely examining it, it has seemed to me that the posterior border of the grey commissure, in place of being suddenly arrested, was diffuse and insensibly confounded with the centre of the posterior columns, as if the latter commenced to be altered in their depth."

Several cases are recorded in which the symptoms of locomotor ataxy occurred, but which recovered entirely. It can scarcely be supposed that in such the nerve fibres had been completely disorganized.

The causes above referred to are calculated to produce derangement in the circulation. The effect of damp cold in deranging the circulation is well known, and the vaso-motor nerves are peculiarly susceptible to influences of an emotional nature.

From these facts in regard to some of the fugitive symptoms of the disease, from the case in which such accurate observers as MM. Gubler, Luys and Duchenne found nothing but congestion of the posterior cords, from the fact that patients have entirely recovered after the second stage of the disease has made great progress, and from the nature of the causes which act to produce it, I am inclined to look upon it as an affection primarily of the vaso-motor nerves, producing congestion, which being only moderate in degree and being long continued causes proliferation of connective tissue, and, as it were, strangles the nerve fibres, causing their absorption.

The question arises, what is the cause of this congestion, of this vaso-motor disturbance? The sympathetic is looked upon as the regulator of the bloodvessels, and disturbance of its function or lesion of its structure interferes materially with the distribution of the blood. M. Duchenne is inclined to refer the changes of the cord to changes in that nerve. In an article in the *Gazette Hebdomadaire*, Feb. 19 and March 4, 1864, he advocates this view. He founds his belief principally on the vascular phenomena connected with the eye, which has been already referred to, and on the symptoms connected especially with the bowels, bladder and other viscera. He thinks that if the ganglia connected with the lower part of the cord were examined, changes might be found to throw light upon the sclerosis of the cord.

One case is recorded by Donnezan in the *Gazette Hebdomadaire*, May 6, 1864, in which, though the autopsy was not complete, a partial answer is given on this point. M. M—— was attacked with the pains of this disease in 1858, and in 1860 was unable to walk without support. Obstinate constipation caused a distressing sensation of a girdle around the body. Towards the last, he lost all motor power, and died in 1864. At the autopsy, the posterior columns in the cervical region were found somewhat affected; the dorsal and lumbar regions were extensively changed. Only the upper cervical sympathetic ganglion could be obtained for examination, owing to the opposition of friends. It was found to be harder, more resisting, and of a more yellowish white than in health. The cells did not differ from those of a healthy ganglion. The communicating branch, however, had become tendinous; the proper nervous tissue had disappeared.

Friedreich, on the other hand, gives a very detailed account of an autopsy, and states that the sympathetic was normal.—*Virchow's Archives*, vol. xxvi.

In an examination made by Luys, and quoted by Trousseau, the ganglia on the posterior roots in the lumbar region were all increased in size, and unusually red and vascular. Their enveloping membrane was considerably thickened. On section, the capillaries were found dilated. The ganglionic corpuscles (cells) were covered with brown pigment-granules, and some were torn and shrunken; others were voluminous and pale. No part of the sympathetic was examined.

So far as I have been able to learn, this is the extent of the investigation in this di-

rection. Much can be said in opposition to the theory of the sympathetic origin of the disease. Nothing can be settled till a larger number of cases have been examined with particular reference to this point.

Treatment.—The benefit to be anticipated depends much upon the view taken of the pathology. If the disease is, in its earlier stages, essentially a vaso-motor disturbance, a simple congestion, the expectation of benefit would be greater the earlier treatment was commenced.

Cases have been recorded where the loss of coördinating power was so great that the patient could not stand; yet in three or four months restoration was complete. It is not necessary, then, to despair of benefiting a patient, even when he seems to be in the advanced stages of the disease.

Electricity seems to do good occasionally, but at best the benefit from it is very uncertain.

The best treatment seems to be, with proper diet and hygiene, that proposed by Wunderlich—nitrate of silver in quarter-grain doses three times a day for four or five weeks; after that time it would be well to omit the drug for a week or so, to avoid coloring the skin. The same course may then be repeated. It may be necessary to give the nitrate in smaller doses at first, on account of gastric irritation, and occasionally it must be entirely omitted. If no benefit follows, the dose should be increased. Althaus gives, with the nitrate, the hypophosphite of soda, and thinks it better than the nitrate alone.

Many have been relieved by this treat-

ment. In only a few cases has complete cure been obtained. One such is to be found recorded in the *Gaz. des Hop.*, Jan. 4, 1863.

Carré mentions 16 cases where a cure was obtained, or the benefit was considerable. In one of these, the nitrate was administered hypodermically, near the spinal column. In 6 cases, the benefit was only partial; either a relapse occurred, or after a while the drug lost its power, or could not be tolerated. In 4 cases, no benefit at all was obtained.

The good effect of the drug is usually observed at the end of a week or fortnight. The first favorable effect is relief of the pain, subsequently return of sensibility, and finally return of coördinating power. Sometimes sight is restored. Simultaneously the general health is benefited, digestion is improved and constipation ceases.

If the congestive theory is correct, bromide of potassium, belladonna and ergot might be of benefit.

NOTE.—It will be noticed that I have used the term "congestion" to express the primary morbid condition. It is considered by many to be a chronic myelitis. Without intending to deny this, I have used "congestion" to express the view that hyperæmia of the part was the cause of the subsequent changes, and to bring out more clearly the rôle played by the vaso-motor nerves. The reason the posterior columns are generally attacked may be that there is a greater abundance of vessels and interstitial tissue naturally in them. See *Archives de Physiologie Normale et Pathologique*, No. 2, p. 329.

